Cardiac Arrhythmias and Sudden Death after Surgery for Congenital Heart Disease

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There has been huge progress in our understanding and management of arrhythmias in patients with congenital heart disease. Our understanding of factors that are associated with arrhythmias have lead to improvements in surgical technique and also to better risk stratification of patients at risk of arrhythmias and sudden death so that therapies can be appropriately targeted. This thesis first addressed the issue of arrhythmias in patients with single ventricles undergoing the Fontan operation, followed by risk stratification for sudden death in patients who had undergone repair of tetralogy of Fallot. Lastly it addressed the issue of risk stratification for sudden cardiac death in patients who have undergone atrial repair for transposition of the great arteries.

Fontan Operation

Chapter two presents an evaluation of the effect of a modification of the Fontan operation on the incidence of and the morbidity/mortality resulting from these arrhythmias. Forty patients undergoing the older operation (atriopulmonary connection (APC) were compared with 40 undergoing the newer operation (total cavopulmonary connection (TCPC). None of the patients had an arrhythmia prior to their surgery. The underlying diagnosis of the form of single ventricle varied between the two groups and indeed suggested that the TCPC group was disadvantaged by the presence of more complex forms of single ventricle compared to the APC group who predominantly had tricuspid atresia which is thought to be one with the best results as far as the Fontan operation is concerned. Despite this, in-hospital mortality was higher (15/40) for the APC and lower (6/40) for the TCPC. Atrial flutter was noted in 9 patients with APC and two with TCPC. More alarmingly, 8 of the 9 APC patients with flutter died. Neither of the two TCPC patients with atrial flutter died. Follow up was short in the study (mean 38 months after APC and 20 months after TCPC) and the incidence of late arrhythmia was low in both groups. Thus this study found a significant improvement in the incidence of early atrial flutter and a significantly lower mortality associated with flutter in the total cavopulmonary connection group. This was a key paper which led to the abandoning of the original APC Fontan and the institution of the TCPC as the favored surgical technique of the Fontan operation.

Since this paper was published, there has been much progress in our understanding of arrhythmias in patients undergoing the Fontan operation. Cecchin et al compared the arrhythmia-free survival of the APC Fontan versus the TCPC Fontan (1). They were able to show a significant reduction in the short and medium term incidence of arrhythmias in the TCPC group. More recently, there has been an interest in avoiding the atrium altogether and performing an extra-cardiac conduit to connect the inferior vena cava to the pulmonary artery (2). Known as the extra-cardiac conduit (ECC) style of Fontan, this modification has been shown to have a lesser incidence of arrhythmia than the intra-cardiac style of TCPC (also known as the intra-cardiac lateral tunnel technique). Two studies, one from Azakie et al (3) and another by Ovroutski et al (4) showed that the ECC was associated with fewer arrhythmias in the short term. Long term studies are still lacking but the ECC has become the technique of choice for the Fontan procedure in most institutions.

C**hapter 3** reviewed the management of patients with Fontan (atriopulmonary connection) having atrial flutter. Eighteen patients referred for management of atrial flutter to a center specializing in the care of arrhythmias were reported. Fifteen of these patients underwent electrophysiologic testing and 12 had sinus node dysfunction. Also, all 15 had inducible atrial flutter at the time of electrophysiologic testing. A variety of management approaches were undertaken. These patients were tried on a variety of anti-arrhythmic drugs (2.7 drugs/patient) with poor results. Anti-tachycardia pacing was done in 16 and were thought to be useful in 8 patients. Radiofrequency catheter ablation was attempted in three patients with one success. Surgical right atriectomy was performed in three patients with benefit in two and one death. One patient underwent His Bundle ablation and pacing. Three of the patients died during follow-up, two suddenly and one after a pacemaker operation. One of the patients underwent heart transplantation. Thus, this paper highlighted the problems and difficulties in the care of Fontan patients having recurrent atrial flutter.

Since this paper was published there has been an enormous amount of effort to improve the treatment of Fontan patients with arrhythmias. Much of our understanding of the macro-reentrant arrhythmias seen in Fontan patients comes from the group in St Louis, Missouri. In a series of elegant papers published in the 1990s, they showed that the incisions placed during the Fontan procedure were themselves arrhythmogenic by allowing reentry around the scars. Gandhi et al showed this to be the case in the classic APC, and that surgical ablation was possible by extension of the scars/creation of new scars inorder to disconnect the reentry circuit (5). Rodefield et al showed the same case to be true for the TCPC (6).

The advent of catheter ablation has provided a significant new approach for this problem. A number of studies have shown that catheter ablation can produce significant acute success in the treatment of this arrhythmia (7-13). However, recurrence rates are still high. The procedures are often complex and time-consuming. Recent improvements in mapping technology, particularly the availability of 3-dimensional electroanatomic mapping has had a significant impact on our understanding of these arrhythmias and our ability to target areas within the atrium responsible for these arrhythmias.

Another approach taken by some authors, notably the group from Children's memorial Hospital in Chicago has been the advent of arrhythmia surgery in Fontan patients. The surgical approach currently involves conversion of the APC to an ECC type Fontan, accompanied by a Maze operation as originally described for adult atrial fibrillation, and implantation of a dual chamber pacemaker (14-16). Significant success in controlling arrhythmia has been reported not just by the Chicago group but others as well (17-19).

Another approach taken by some has been to create prophylactic incisions within the atrium to prevent the development of arrhythmias. Preliminary studies of this aspect by Collins et al have yielded unconvincing results (20).

Chapter 4 deals with the area of pacing in Fontan patients. There is a significant long term risk of bradyarrhythmia with need for pacing in this group. There has been no study of the effect of the type of pacing (atrium alone versus ventricle alone versus both chambers) in this patient group. This prospective study was an acute cross-over trial performed in the intensive care unit soon after the Fontan operation and studied 21 patients aged 2 to 18 years (median 4). Ventricular-only pacing showed significant worsening in hemodynamics with lower cardiac output, a higher left atrial pressure and higher pulmonary arterial pressure and lower systemic blood pressure. No significant differences were noted between atrial only versus dual chamber (atrial and ventricular) pacing. To our knowledge this is the only systematic study of this important problem.

Tetralogy of Fallot – Ventricular Tachycardia and Sudden Death

Chapter 5 reported on the correlation between prolonged QRS duration and the occurrence of sustained ventricular tachycardia at electrophysiologic study in a cohort of 135 patients who had undergone repair of tetralogy of Fallot. Sustained ventricular tachycardia was induced in 22 patients. Induced sustained ventricular tachycardia was associated with prolonged QRS duration. A QRS duration >180 milliseconds was 35% sensitive and 97% specific for association with induced sustained monomorphic ventricular tachycardia and was 100% sensitive and 96% specific for patients with clinical ventricular tachycardia (i.e. ventricular tachycardia noted in the real world circumstance and not just induced in the electrophysiology laboratory). This paper therefore argued for the intensive evaluation and follow-up of patients with QRS duration >180 milliseconds even if they were asymptomatic.

Chapter 6 is a report on risk stratification for sudden death in patients with tetralogy of Fallot. In a multi-institutional study, a total of 793 patients were recruited from six international centers. Of the 793, 33 developed ventricular tachycardia, 16 died suddenly and 29 had atrial arrhythmias (atrial flutter or fibrillation). A prolonged QRS duration and accelerated rate of change in QRS duration over a ten-year period were found to be associated with ventricular tachycardia and sudden death. Patients with sudden death or atrial tachyarrhythmias had undergone surgery later in life. Also the study showed that pulmonary regurgitation was the main underlying hemodynamic problem in patients with ventricular tachycardia and sudden death, whereas tricuspid regurgitation was the main finding in patients with atrial flutter or atrial fibrillation. Thus this paper further bolstered existing views about the intimate link between poor hemodynamics and late arrhythmias in this group of patients. It offered a rationale for pulmonary valve surgery to restore competence in patients as a strategy to decrease the incidence of sudden death.

Transposition of the Great Arteries Status Post Atrial Switch Procedure

Chapter 7 reported a study on predictors of sudden cardiac death in patients who have undergone atrial repair (Mustards/Sennings operation) for transposition of the great arteries. In many prior studies sudden death was the predominant cause of late death. This was a retrospective case-controlled study of 47 patients with sudden death (or near-miss sudden death) and two controls per patient who had not died suddenly. It looked at

clinical parameters from the patient such as symptoms by history, physical examination findings, ECG findings, chest Xray, echocardiogram and 24-hour ambulatory ECG findings. This study noted the following: symptoms of arrhythmia or congestive heart failure at follow-up and a history of documented arrhythmia (atrial flutter/atrial fibrillation) were associated with sudden death. Other parameters such as electrocardiogram, chest x-ray and holter electrocardiogram were not predictive of sudden death. Neither medical (drug) therapy nor pacing was shown to have a protective effect for sudden death. Most sudden death events occurred during exercise and ventricular tachycardia or ventricular fibrillation were the final recorded event in 21 of 47 patients. Thus, beyond identifying predictors, this paper also emphasized the importance of exercise restriction in patients at risk for sudden death. This paper also raised the possibility that successful catheter ablation of atrial arrhythmias may lower the risk of sudden death in these patients.

Conclusion

In conclusion, this thesis highlights the role of arrhythmias andsudden death after surgery for congenital heart disease and outlines approaches to prevention (using surgical modification for example), and risk stratification for sudden death.

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